## 281 Cross-sectional- and long-term-ultrasound bone density assessment in pediatric and adult patients with Cystic Fibrosis

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Bone density assessment regularly implies radiation and often lacks of regional normative values, especially in childhood.

Aim of the present study was to assess ultrasonic bone density (BD) at the calcaneal bone from pediatric and adult patients with CF in a cross-sectional- and long term setting. The pediatric results were compared to regional reference data obtained with the identical system from 3299 healthy controls aged 6 to 18 yrs. BD parameters were assessed with the SAHARA$^{TM}$ system in 54 CF patients in a cross sectional setting and in 35 CF patients for a long term of up to 7 years.

Cross sectional analysis revealed an increased rate of reduced BUA and SOS-values in CF patients [BUA $\leqslant$ 1 standard deviation score (SDS): 13/54 (24.1%) resp. >1 SDS: 7 (13.0%); SOS $\leqslant$ 1 SDS 16/54 (29.6%) resp. >1 SDS: 10 (18.5%)]. The differences were more pronounced in children than in adults. Long term observation revealed a decrease of BUA and SOS Z-scores in the 21 patients followed for at least 48 months. Most distinctive reduction was found in SOS Z-scores from children controlled after 12–24 months ($n=7$, mean $=-0.47$, $p=0.003$) and in adults controlled after 24–36 months ($n=20$, mean $=-0.39$, $p=0.032$).

Altogether we found a relevant proportion of reduced BUA and SOS values in CF-patients. The higher proportion of children with reduced values could coincide with the presence of relevant local reference data, whereas we did not propose about similar references in adults.

Although variance of ultrasonic BD-measurement is higher than in the standards DXA or Q-CT, which imply radiation, the method may be a tool for frequent follow-ups in CF.

## 282 Frequency and risk factors of osteopenia/osteoporosis in children with Cystic Fibrosis

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It was shown in different studies that reduced bone mineral density (BMD) is often complication in CF and can significantly reduce life quality of these patients.

**Aim:** to determine frequency of osteopenia and osteoporosis in CF children, to compare frequency of reduced BMD in CF children and healthy Russian population the same age and to identify risk factors.

**Methods:** 44 CF children (24M/20F, 5.4–17.9 yrs, mean FEV1% 67.87%±23.14) were studied. 14 (32%) children have "moderate" and 30 (68%) "severe" phenotype. 5 (11%) children used inhaled steroids, 7 (16%) – oral steroids (0.5 mg/kg more than 1 year). Bone mineral content(BMC) and BMD are measured using DEXA scans of the lumbar spine (L2-L4). Anthropometric parameters, clinical markers of disease severity, pulmonary function tests (FVC, FEV1%), serum calcium and phosphor, alkaline phosphatase were assessed and related to bone mineral density.

**Results:** osteoporosis was found in 43% (meanZ-score-3.5, S.D. 0.97), osteopenia (mean Z-score-1.52, S.D. 0.35) in 34% cases. However the complaints on a pain in bones and back were only at 7 (16%) patients, nobody had an atraumatic fracture. While at healthy Russian children low BMD is revealed at 10–30%. There was no correlation between BMD and gender, age, inhaled steroids. We have observed correlation between BMD and weight/height ($r=0.38$,$p<0.05$), FEV1% ($r=0.39$,$p<0.02$), FVC ($r=0.39$, $p<0.02$), long term oral steroids ($r=0.41$,$p<0.01$).

**Conclusion:** reduced bone mineral density is often complication in CF children. The frequency reduced BMD at children with CF in 2 times is higher than at healthy children. The condition of lungs functions is one of the important factors determining BMD.

## 283 Prevalence of osteopenia/osteoporosis in an adult CF centre

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Bone loss is a recognised complication of CF. Previously we performed DEXA scans on patients we identified as being at increased risk of developing bone disease:(steroids, low Vit D levels, low BMI, diabetes, >40 years, on transplant list). Results confirmed a high prevalence of bone disease in this group. We performed DEXA scans on the remaining clinic population to determine overall prevalence in our clinic.

**Aims:** To investigate the prevalence of bone disease in all patients in our CF centre. To review the current treatment of bone disease in our clinic population. To investigate relevance of repeating DEXA scans 2 yearly. To update guidelines for the detection and management of bone disease within our CF centre.

**Methods:** DEXA scans were performed on all patients in our clinic. Patients who had previously been scanned had a repeat scan 2 years after their initial scan.

**Results:** DEXA scans have been performed on 98 patients. 23 have osteoporosis, 47 have osteopenia and 12 have normal scans. 16 patients were <20 years old and could not be classified under the WHO definition. 27 repeat scans have been performed with 4 results showing different diagnosis from original scans. 3 results changed from osteopenia to osteoporosis, 1 result showed change from osteopenia to normal. Further analysis of data is ongoing to look at risk of low vitamin D levels within clinic.

**Conclusions:** Bone loss is high in our CF population, including in some patients identified as low risk. Results in those <20 years are difficult to interpret as bone is still being laid down. This raises questions as to when treatment should be instigated in this group. Changes in bone density can be detected within a 2-year period. Guidelines require to be updated to include treatment for our <20 years population.

## 284 Reduced bone density in patients with Cystic Fibrosis in the Republic of Macedonia

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The aim of the study was to determine the prevalence and identify determinants of reduced bone mineral density (BMD) in children and adolescents with cystic fibrosis (CF).

Children (30) with CF (mean±SD age 11.5±4.5 yrs); forced expiratory volume in one second (FEV1) 87.8±24.5% of the predicted value was studied.

The method of quantitative ultrasound (QUS) was used for determination of the bone status. Speed of sound (SOS), transmission index (TI) and osteosono-assesment (OSI) were measured by calcaneal ultrasound.FEV1 (% pred); CF genotype; malnutrition, history of growth, development or weight gain delay and corticosteroids use were analysed.

The mean values of Z score in CF patients were for SOS (–0.2±1.4); for TI (–0.6±1.1) and for OSI (–0.5±–1.2). We found 10 patients (33%) with reduced bone density. All of them were homozygous or heterozygous for the ΔF508 mutation. Malnutrition, lower 25-hydroxyvitamin D level and lower FEV1 (% pred) associated with lower BMD.

In conclusion, reduced BMD in CF patients is associated with a number of factors, including ΔF508 genotype, male sex, greater lung disease severity and malnutrition